

Use of new oral anticoagulants and pulmonary thromboembolism recurrence secondary to Paget–Schroetter syndrome: A case report

Morales MC, Delgado AFl, Lemus HN, Montenegro AC

ABSTRACT

Introduction: The ‘effort’ thrombosis or Paget–Schroetter syndrome is an unusual form of upper limb deep venous thrombosis. The current management of this syndrome consists of surgical approach and to this date, there is no evidence of the anticoagulant therapy effectiveness. **Case Report:** We describe a case of a young Korean male with pulmonary embolism secondary to Paget–Schroetter syndrome. The patient was treated with rivaroxaban for pulmonary embolism and no surgical intervention was done. After a follow-up at 6th and 12th months, there was no recurrence of any thrombotic event. **Conclusion:** We described a clinical scenario in which the use of new oral anticoagulants for the treatment of a pulmonary embolism prevents the recurrence of an upper limb thrombosis secondary to Paget–Schroetter syndrome. Even though there is no evidence in favor of using new oral anticoagulants for upper limb thrombosis, the current report proves its successful use.

Keywords: Effort thrombosis, New oral anti-coagulants, Pulmonary embolism, Upper limb thrombosis

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INTRODUCTION

Paget–Schroetter syndrome or ‘effort’ thrombosis is an unusual form of upper limb deep venous thrombosis. It affects predominately young people with an inherent anatomic variant at the costoclavicular junction that triggers the formation of a primary thrombosis [1]. It is important to suspect and identify this syndrome in order to prevent further complications, such as pulmonary embolism (PE), post-thrombotic syndrome, or even death. Herein, we present a clinical case in which a patient develops a PE secondary to an effort thrombosis and was managed with a new oral anticoagulant.

CASE REPORT

We describe the case of an 18-year-old male, from South Korea, who presented to the emergency department with one-week course of left upper limb pain with swelling and mild erythema. There was no previous history of chest trauma and patient denied having fever. These symptoms progressed in the last three days prior to the hospital admission. The patient took 100 mg of aspirin without any improvement of the symptoms. Furthermore,

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the patient referred having an episode of acute dyspnea and pleuritic chest pain the previous night. The patient had no known medical problems, no medications and no prior family history of hematologic-related diseases. Of note, the patient is a golf and volleyball player. On physical examination his blood pressure was 106/68 mmHg, pulse rate 114/min, pulse oximetry showed 92% on room air, and temperature was 36.4°C. His lungs were clear on auscultation and his heart sounds were normal. Edema, tenderness and erythema were found over the left upper extremity, distal perfusion of one second and no motor or sensitive deficit was present. He did not have Urschel's sign.

With the previous findings we decided to perform a duplex ultrasonography that showed a thrombosis from the middle third of the left subclavian vein to the innominate vein. The rest exams were normal. We initiated anticoagulation with low molecular weight heparin (LMWH) enoxaparin at therapeutic doses (1 mg/kg twice a day). According to the clinical presentation and pretest probability for PE (Wells score) we decided to perform a computed tomography scan angiography (Figure 1). It confirmed the diagnosis of PE on the base-lateral segment artery of the right inferior pulmonary lobule. An echocardiogram ruled out a right heart compromise.

In the second day of hospitalization, the patient presented a satisfactory evolution and was discharged on oral anticoagulation with rivaroxaban 15 mg twice a day for the first 21 days, and then 20 mg per day to complete six months; this with the aim to treat PE.

In contrast to the algorithm described by Allan [2], for the treatment of Paget–Schroetter syndrome, we focus the patient's treatment towards PE as the main short-term morbi-mortality cause in this patient.

The patient was followed-up at 6th and 12th months later, with no evidence of recurrence neither PE nor upper limb thrombosis. In addition, he did not required surgical intervention, and has been playing golf since we saw him during hospitalization.

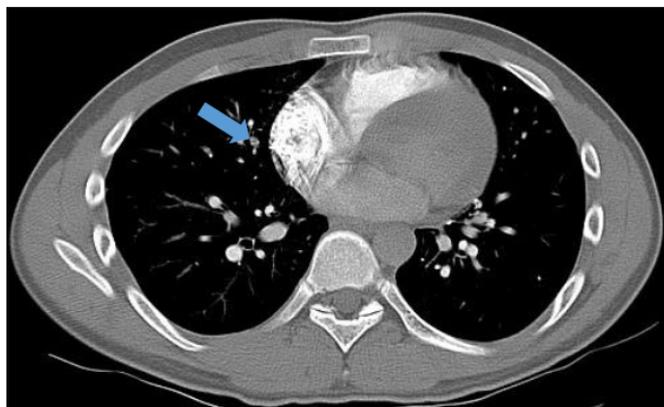


Figure 1: Computed tomography angiography: blue arrow indicate thrombus in the artery for lateral-basal segment.

DISCUSSION

The Paget–Schroetter syndrome or “effort thrombosis” is one of the venous thoracic outlet syndromes characterized by a primary thrombosis in the subclavian vein at the costoclavicular junction [1].

The Paget-Schroetter syndrome is a rare condition, with an estimated incidence of 1 per 100,000 population and represents 1–4% of the total events on venous thrombosis. However, the incidence seems to be underestimated because of the poor clinical identification. It was reported that Paget–Schroetter syndrome accounts 30–40% of spontaneous axillary-subclavian vein thrombosis and approximately 15% of all upper limb thrombosis [3]. The prevalence of right-hand dominance explains why thrombosis is more frequent in the right subclavian vein. Furthermore, is more frequent in men with an estimate ratio of 2:1 [1].

The pathophysiology of this syndrome requires of a good anatomical understanding of the superior thoracic outlet region, especially on the anterior compartment. Behind the scalene fat lies the anterior scalene muscle from the cervical spine vertebrae to the top of the anterior first rib. The middle scalene muscle lies from the cervical spine vertebrae to the mid segment of the anterior first rib. The space between these two muscles is known as the scalene triangle. Structures such as the brachial plexus and subclavian artery are found in the scalene triangle, meanwhile, the subclavian vein lies in front of the anterior scalene muscle in the intersection of the clavicle and first rib [2].

Two theories explain the initial process that generates the final event of thrombosis in the Paget-Schroetter syndrome. The first one involves frequently activity of the arm followed by hypertrophy of the anterior scalene muscle; this, leading to blood stasis and clot formation. Secondly, several descriptions suggest that secondary thrombosis begins with a structural compression (anatomic abnormalities in the upper thoracic outlet), which triggers a continuous process of fibrosis and scarring in the external environment surrounding the subclavian vein. These perpetuate blood stasis and thrombosis [4].

The typical clinical presentation is a sudden onset pain, heaviness, blue-red discoloration and swelling of the upper affected arm. However, due to the mainly presentation in young athletic patients, the syndrome tends to be asymptomatic, or described by the patient as a simple muscular strain. In patients with intermittent obstruction, the symptoms will appear and disappear according to the process of obstruction. Occasionally, patients present the ‘Urschel's sign’ characterized by the dilation of veins that can be visible across the shoulder and upper arm [2].

Diagnosis is based in clinical signs, symptoms and medical records. Duplex ultrasound scan is the best diagnostic test [5]. The sensitivity is found to be 78–100% and the specificity 82–100% [6]. A recent clot will be echolucent, whereas more chronic clots are associated

with an echogenic appearance. The absent flow and lack of compressibility can establish the diagnosis. In addition, the visualization of collateral venous pathways can enhance the probability of diagnosis.

The mainstream treatment of upper extremity thrombosis is based on the correction of the underlying defect and prevention future episodes. Currently, the gold standard in the acute management of this unusual thrombosis is the thrombolytic therapy. Catheter-directed thrombolysis has reported a successful of 62–82%, being higher in recent formed clots (symptoms <2–6 weeks) followed by a venoplasty if there is evidence of residual obstruction [2, 7]. Anticoagulation therapy reduces overall mortality and morbidity associated with this syndrome. However, some studies demonstrate an increased risk of residual venous obstruction in 78% of cases managed only with anticoagulation [7]. Urschel et al. describe that only 29% of patients treated with anticoagulation reported a good or excellent outcome [8]. There was no evidence related to the use of new oral anticoagulants for the management of upper limb thrombosis.

In cases of external compression by scarring and fibrosis the treatment includes surgical decompression to avoid subsequently new thrombosis [9]. The surgical procedure should be performing one or two months after the episode.

CONCLUSION

Paget–Schroetter syndrome is an upper limb thrombosis related to young athletic patients. Its pathophysiology is explained by two theories: an overuse of the extremity with adnexal muscle hypertrophy and intrinsic anatomical abnormalities in the thoracic upper outlet; both enhance the probability of blood stasis and thrombosis. The current treatment for this syndrome is the correction of any anatomic abnormality with surgical interventions. There are no studies related with the use of new oral anticoagulants for its management. In this case report, we have shown how rivaroxaban was used successfully for the treatment of pulmonary embolism and also prevents the recurrence of the upper limb thrombosis. Further clinical trials are needed to show the clinical benefits of this drug and prove an association.

Author Contributions

Morales MC – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Delgado AFL – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Lemus HN – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Montenegro AC – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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